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Choledochal Cyst Disease

A Changing Pattern of Presentation

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Objective
The authors compared the presentation, treatment, and long-term outcome of children and adults with choledochal cysts.

Summary Background Data
The typical patient with choledochal cyst disease has been the female infant with the triad of jaundice, an abdominal mass, and pain. However, the recent experience of the authors suggested that the disease currently is recognized more commonly in adults.

Methods
Forty-two patients (11 children, 32 adults) with choledochal cyst disease were treated primarily at this institution between 1976 and 1993. Patient presentation, clinical evaluation, and operative treatment were obtained from existing records. Long-term follow-up was obtained by records, physician, or direct patient contact.

Results
One child—but no adults—had the classic triad of jaundice, abdominal mass, and pain. Children were more likely to have two of the three signs or symptoms (82% *vs.* 25%; $p = < 0.05$). Adult patients most commonly had abdominal pain and were thought to have pancreatitis (23%) or acute biliary tract symptoms, prompting cholecystectomy (50%). The type of choledochal cyst seen in children and adults was similar; the fusiform extrahepatic (Type I) was most common (50%), and the combined intrahepatic and extrahepatic (Type IVA) was the next most prominent (33%). For both children and adults, treatment consisted of excision of the cyst and biliary reconstruction with a hepaticojejunostomy. There was no surgical mortality. Gallbladder or cholangiocarcinoma was identified in three adults (9.7%), two of which were manifest on presentation. Long-term follow-up revealed one patient with a biliary stricture and three patients with Type IVA cysts who had intrahepatic stones.

Conclusions
Children and adults differ in presentation of choledochal cysts, with adults commonly having acute biliary tract or pancreatic symptoms. Surgical treatment with cyst excision and biliary bypass is safe and effective in children and adults with excellent long-term results that minimize the development of malignancy.

Choledochal cysts originally were described in 1723 by Vater and Ezler.[1] The first surgical excision of a choledochal cyst was performed by MacWorter in 1924.[2] Over 3300 cases of choledochal cyst disease have been reported; more than 1800 of these cases have been compiled from the Japanese literature alone.[3] In the United States, choledochal cysts remain uncommon, and most reports of this disease outside of Asia contain small numbers of patients.[4,5] The typical patient with choledochal cyst disease has been a female infant or child with the triad of jaundice, abdominal pain, and a palpable mass. Recently, however, at The Johns Hopkins Hospital, the diagnosis of choledochal cyst disease has occurred more commonly in adults than in children. Therefore, we reviewed our experience with patients with choledochal cyst disease, emphasizing the differences in presentation and pathology between children and adults.

CLINICAL MATERIAL

The medical records of all patients with the diagnosis of choledochal cyst disease treated at The Johns Hopkins Hospital between 1976 and 1993 were reviewed. Presenting complaints, physical examination, preoperative evaluation, and treatment were recorded. Ultrasound examinations, computed tomography scans, cholangiograms, and operative reports were reviewed to determine the type of cysts. Cysts were classified according to the Todani modification[6] of the Alonso-Lej classification (Fig. 1). Material from resected cysts and liver biopsies were reviewed by a pathologist (JKB). Four patients treated elsewhere with choledochal cyst excision and referred for subsequent anastomotic strictures were excluded from this analysis. Long-term follow-up of patients was obtained from medical records, primary physicians, or direct patient telephone communication. Of the 42 patients, one child and one adult were could not be observed for follow-up. Differences between children and adults were compared by chi square or t test for unpaired data, as appropriate.

Patient Demographics

Forty-two patients (31 adults, 11 children) with choledochal cyst disease were analyzed. One of the children was considered an adult for cyst excision. Thus, 43 different presentations of choledochal cyst disease occurred in the 42 patients. Thirty-six patients were women, and six were men. Patients 16 years of age or younger were characterized as children. Age at diagnosis ranged from antenatal to 68 years. However, 9 of 11 children were identified by 4 years of age or younger (median age 15 months). Adult patients ranged in age from 17 to 68 years, with a median of 30 years. In the first 8 years, 1976 to 1984, only ten patients with choledochal cysts were seen—five children and five adults. However, from 1985 to 1993, 32 patients with choledochal cyst disease were treated, including 6 children and 26 adults.

Presenting Symptoms

Only 1 of 11 children had the classic triad of right upper quadrant mass, abdominal pain, and jaundice; none of the adults had this triad. Eight additional children had two of the triad symptoms; only 8 of 32 adults had two of the three symptoms. Thus, two or three classic signs or symptoms of choledochal cyst disease were significantly more common in children than adults (82% *vs.* 25%; $p < 0.05$). On the other hand, 7 adult patients (23%) had signs and symptoms of pancreatitis, and 16 adult patients were thought to have symptomatic gallstones or acute cholecystitis and had been explored previously for suspected gallbladder pathology. One adult had biliary tract symptoms and was found to have a gallbladder carcinoma at cholecystectomy. Pancreatic or gallbladder pathology was not suspected primarily in any child. Table 1 demonstrates the presenting symptoms and signs in children and adults. As might be expected, the time from the onset of symptoms until diagnosis was significantly shorter for children than adults (8 months *vs.* 6 years; $p < 0.01$).

Preoperative Evaluation

Elevated bilirubin, alkaline phosphatase, transaminases, or serum amylase levels were seen in 31 of 42 patients (72%). Liver function test abnormalities were present in all children and 20 adults (63%). Elevated serum amylase levels were not seen in children but were elevated before operation in 12 adults (38%, $p < 0.05$). Similarly, clinical pancreatitis was present in none of the children and in seven of the adults with elevated amylase levels (22%, $p = 0.06$).

The diagnosis of choledochal cyst disease was established in both children and adults with a combination of ultrasonography (27 patients), computerized tomography (10 patients), and cholangiography, via the percutaneous transhepatic (27 patients), endoscopic (17 patients), or operative routes (6 patients). Many patients had both endoscopic and transhepatic cholangiography. The purpose of using both procedures was to better define the extent of intrahepatic disease (7 patients) or to place a preoperative stent (2 patients). Table 2 depicts

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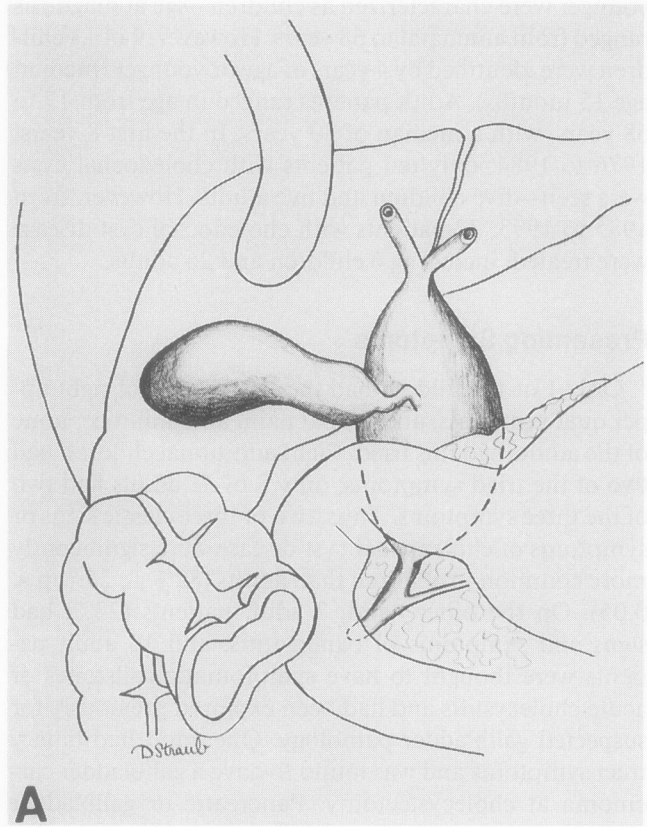
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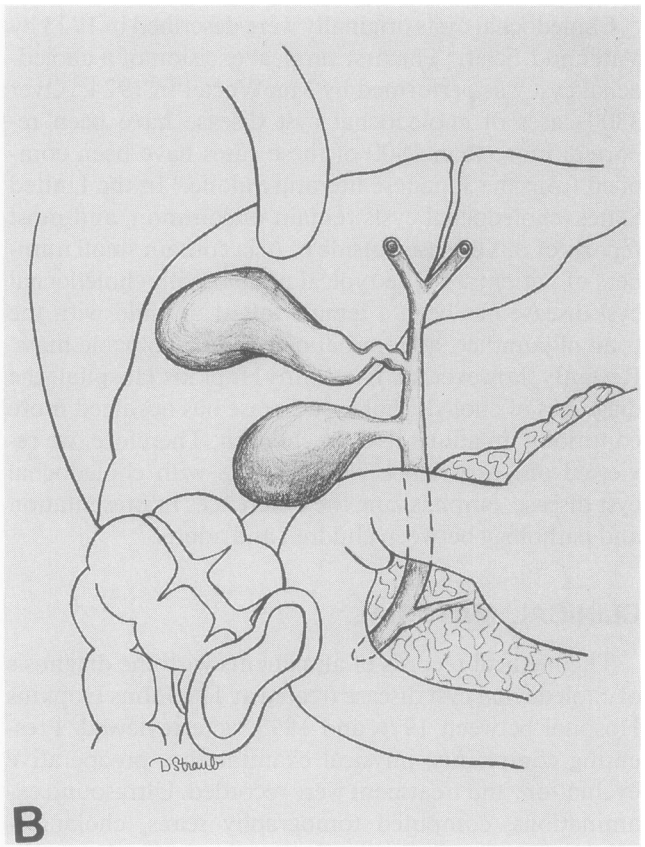
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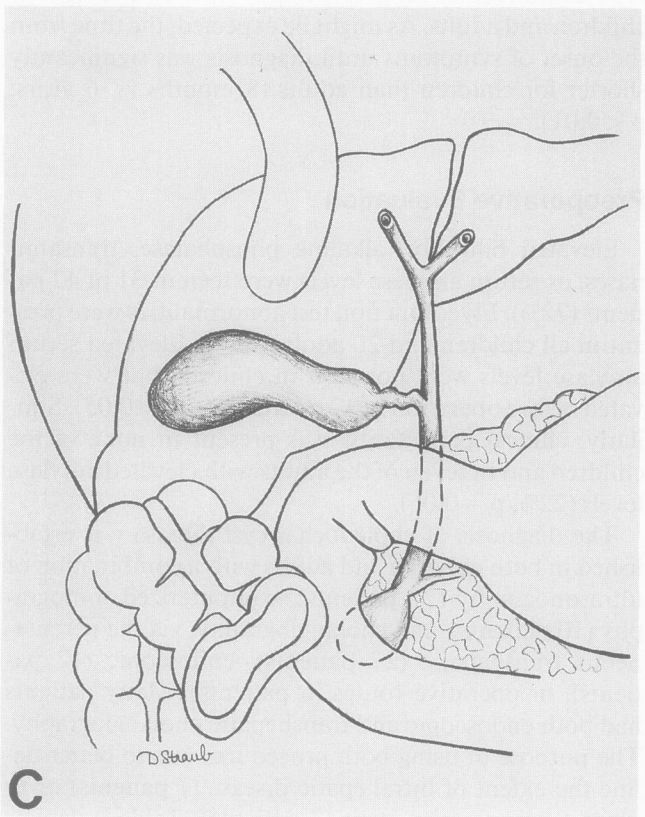
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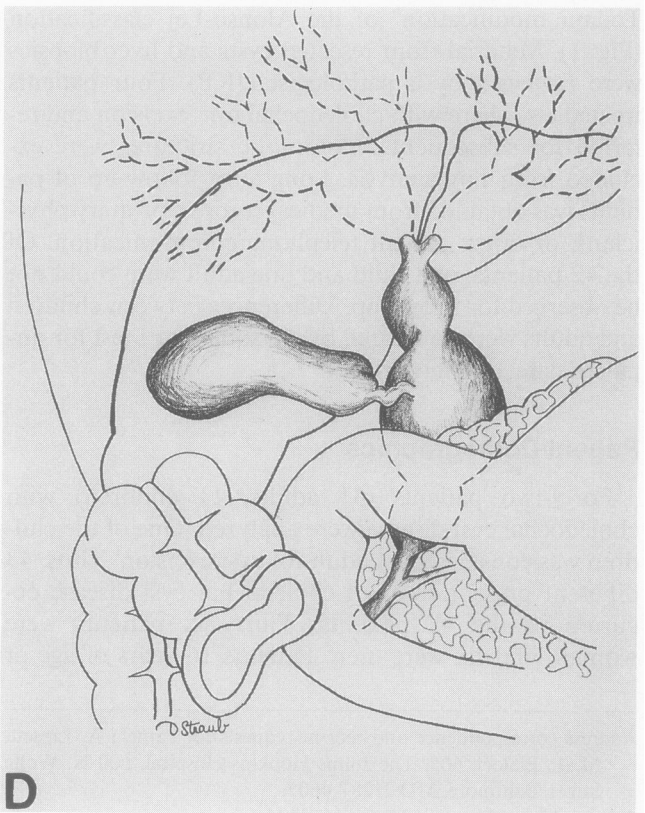
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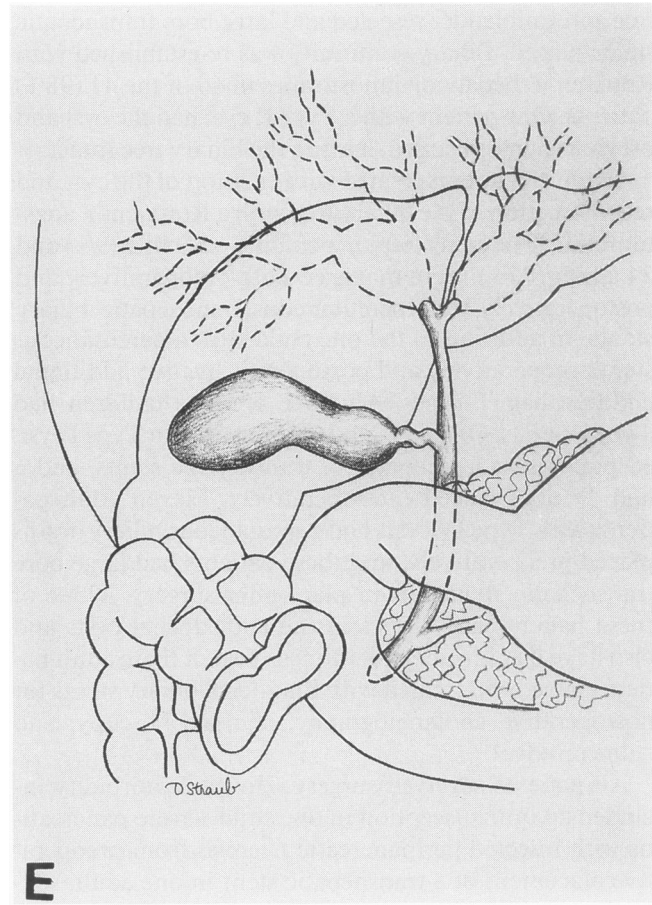

Figure 1. Classification of choledochal cysts as proposed by Alonso-Lej[27] and modified by Todani.[6] (A) Type I: Dilatation of the extrahepatic biliary tree Ia-cystic (shown), Ib-focal, or Ic-fusiform, (B) Type II: Saccular diverticulum of extrahepatic bile duct. (C) Type III: Bile duct dilatation within the within the duodenum (choledochocele) (D) Type IVA: Dilatation of the intrahepatic and extrahepatic biliary tree (E) Type V: Intrahepatic cysts (Caroli's disease).

the results of the diagnostic studies. At Johns Hopkins, ultrasonography, computerized tomography, and cholangiography all were helpful studies. Percutaneous cholangiography and computed tomography scans were most helpful in defining the intrahepatic extent of disease if endoscopic cholangiography was unable to fill the proximal extent of the biliary tree. Nuclear medicine scans often were not diagnostic and required additional confirmatory evaluations, such as ultrasonography, computed tomography scans, and, ultimately, cholangiography.

Cyst Type

The distribution of choledochal cyst types in children and adults is shown in Figure 2. Of the 11 children, Type I cysts were seen in five patients (45%), whereas Type IV cysts were seen in six (54%). Five of these six children had Type IVA cysts. No child had a Type II, III, or V cyst. Of the 31 adults, 17 patients had Type I cysts (55%), and 11 patients had Type IV cysts (35%). Nine of the 11 adult Type IV cysts were subtype IVA. One adult had a Type II cyst (3%), two had Type III cysts (7%), and one had Caroli's disease (Type V). An abnormal choledochopancreatic duct junction (CDPDJ) could be seen in all 15 patients in whom the anatomy was documented. A long common channel was present if the junction of the pancreatic and common duct was longer than 10 mm. Often this junction was at right angles. Of the 15 patients in whom the anatomy could be delineated, two were children. A long and abnormal CDPDJ common channel was identified in patients who had Type I, II, and IV cysts (Fig. 3). In many patients, the size of the cyst obscured clear radiographic visualization of the CDPDJ. No patient in whom the CDPDJ was visualized had a normal pancreaticobiliary junction.

CLINICAL COURSE

Nine of 11 children (82%) and all 30 adult patients (100%) with extrahepatic cysts were treated with cyst excision. One of the adult patients previously had been treated as a child at Johns Hopkins with cyst bypass. The patient with Caroli's disease had his extrahepatic biliary

tree and gallbladder resected and large bore transhepatic stents placed. Biliary continuity was re-established with Roux-en-Y hepaticojejunostomies in 40 of the 41 (98%) patients. One patient with a Type II cyst had the cyst and previous bypass excised, leaving the biliary tree intact.

Of the 40 patients treated with excision of the cyst and reconstruction of the biliary tree with a Roux-en-Y anastomosis, 25 patients (60%), including one child (9%) and 24 adults (77%), were managed both preoperatively and postoperatively with percutaneous transhepatic biliary stents. In addition to the one child with a percutaneous stent preoperatively and postoperatively, two additional children had T tubes. All three of these children had Type IV cysts. Of the 16 adult patients with a Type I cyst, 10 patients had preoperative transhepatic stents, and 6 had T tubes placed intraoperatively. Eleven adult patients with Type IV cysts had percutaneous biliary drains placed preoperatively, and these patients had large bore transhepatic stents left in place after surgery. Three of these patients had stones in the choledochal cysts and also have had intrahepatic stones. Two of these adult patients have been treated with long-term biliary stents for postoperative cholangiography, choledochoscopy, and stone removal.

All patients survived surgery. Hospital morbidity included an intussusception in one child, severe pancreatitis with infected peripancreatic necrosis from preoperative placement of a transhepatic stent in one adult, self-limited pancreatitis in two other adult patients, and a limited bile leak (5 days) in one adult.

Pathology

All children had liver biopsies performed either before or concurrent with cyst excision. Histopathology demonstrated some degree of portal fibrosis, but no established cirrhosis, in these children. The choledochal cyst wall was densely fibrotic in five children with evidence of acute and chronic inflammation in the remaining six children (Fig. 4A). No child had metaplasia, dysplasia, or carcinoma identified within a choledochal cyst. The one adult patient with Caroli's disease developed cholangiocarcinoma within the cysts after a bypass procedure. Severe metaplasia was seen in three additional patients (Fig. 4B). In addition, one adult patient had gallbladder cancer at presentation and diagnosis of her choledochal cyst. Severe inflammatory changes with erosions also were seen in four adult patients, whereas mild acute and chronic inflammatory changes were noted in the remaining 26 adults. Unlike the histopathology seen in children, a dense fibrotic cyst wall was seen in only three adults, with a lesser degree of fibrosis observed in three additional patients.

Follow-Up

Follow-up was complete in 10 children and 30 adults (97%). The median length of follow-up in children was 10 years (range 1–14 years), whereas the median length of follow-up for adults was 3 years (1 month–12 years). No child has developed cholangiocarcinoma during follow-up. In one child, a chronic undiagnosed encephalopathy has occurred, but no hepatic or biliary tract problems have been identified. In the adult patients, long-term morbidity included recurrent cholangitis in three patients. Cholangitis was related to anastomotic stricture formation in one Type IV patient who has been treated successfully with percutaneous dilatation. The two other Type IV patients with cholangitis have had recurrent intrahepatic stones. One of these patients required a left hepatic lobectomy, and the other, who had severe hepatic fibrosis and portal hypertension, was treated successfully with hepatic transplantation. Two additional Type IV adult patients have been managed with long-term stents because of the formation of intrahepatic stones, but they have not been ill. The one adult patient with Caroli's disease who developed cholangiocarcinoma died of metastatic disease.

DISCUSSION

The estimated incidence of choledochal cyst disease ranges from 1 in 13,000 to 1 in 2 million patients.[3,6] The disease is three to four times more common in women than men. More than one half of the reported cases have occurred in Japanese patients. Moreover, the majority of patients in whom it has been reported have been reported are children. In recent years, however, choledochal cyst disease has been reported in increasing numbers of adult patients.[7] In this series, 31 of 42 patients (74%) who presented for treatment were older than 16 years of age. Ten patients had choledochal cysts treated during the years 1976 to 1984; 32 patients were identified during the 1984 to 1993 time interval. During the early time period, the child to adult ratio was 1:1; more recently, the disease has been identified four times more commonly in adults. Furthermore, 16 of 31 adult patients (52%) were thought to have uncomplicated biliary tract disease at the time of initial presentation.

Depending on the patient's age, either an abdominal mass or abdominal pain was the most common presenting symptom. In this series, 82% of the children had abdominal masses, a finding that is similar to the 62% reported by Joseph et al.[8] Furthermore, the series by Lopez and colleagues[9] documents that abdominal pain is a prominent presenting complaint in adults, which also is similar to this review. Moreover, jaundice also was more common in children in the report by Lopez et al.[9] as well

Table 1. SYMPTOMS AND SIGNS AT PRESENTATION

	Children (N = 11)	Adults (N = 31)
	N (%)	N (%)
Abdominal mass	9 (82)	4 (13)*
Abdominal pain	4 (36)	27 (87)*
Jaundice	7 (64)	13 (42)
Fever	2 (18)	8 (26)
Nausea/vomiting	2 (18)	9 (29)
Pancreatitis	0 (0)	7 (23)†
Prior cholecystectomy	0 (0)	16 (52)*

* $p < 0.01$ *vs.* children, chi square.
† $p = 0.06$ *vs.* children, chi square.

Table 2. DIAGNOSTIC EVALUATION OF CHOLEDOCHAL CYSTS

	N	Diagnosis Established
Investigation		
Ultrasonography	27	27 (100%)
Computerized tomography	10	4 (23%)
Cyst type identified		
Cholangiography		
Operative	6	6 (100%)
Endoscopic	17	11 (65%)
Percutaneous	27	27 (100%)

as in this series. The classic triad of jaundice, right upper quadrant pain, and a mass was present in only one child in this series, but children were much more likely to have two of the three classic symptoms.

Although laboratory evaluation may demonstrate mildly abnormal liver function tests or amylase values, these findings are not specific. Ultrasonography, computerized tomography, and cholangiography all are effective in defining the presence of biliary dilatation.[10,11] Ultrasonography and computerized tomography done at this institution always were diagnostic for the presence of a choledochal cyst. However, the large percentage of adult patients with choledochal cysts identified first at the time of cholecystectomy for biliary tract symptoms suggests that ultrasonography may underestimate this diagnosis. The choledochal cyst(s) may have been missed on ultrasonography because of the technical quality of the examination or failure to recognize the unusual pathology. Nonetheless, if choledochal cyst disease is expected, an ultrasound should be diagnostic. However, in an older patient with unusual symptoms relating to the right upper quadrant, or pancreatitis, particularly if gallstones are absent, computed tomography can provide important information about the extrahepatic or intrahepatic extent of biliary dilatation. Although hepatobiliary scintigraphy has been used to diagnose choledochal cysts,[11] we feel that scintigraphy does not provide accurate anatomic detail and, therefore, should not have a role in diagnosing this disease.

Cholangiography is essential in differentiating the type of biliary cyst and in planning the extent of operative resection. Endoscopic retrograde cholangiopancreatography best visualizes the pancreaticobiliary junction, but may not define the superior intrahepatic extent of the cysts if the cysts are redundant and sequester large amounts of contrast material. Preoperative percutaneous cholangiography has been our preferred mode of cholangiography. The advantage of this method has been the ability to define the proximal extent of biliary dilatation and to use this information in the preoperative plan for resection. In addition, we believe that the preoperative placement of a percutaneous ring catheter after cholangiography is of value to the surgeon during the operative procedure and can be used as a stent for the biliary reconstruction. In patients with proximal hepatic duct dilatation (Type IV and V cysts), the preoperative placement of bilateral (right and left) stents has facilitated intraoperative resection and reconstruction of the biliary tree. In several patients long-term postoperative biliary stenting with large bore catheters has allowed access for the management of intrahepatic stones that can form in patients with large intrahepatic biliary sacculations.

Pancreatitis was the only complication of preoperative transhepatic stenting in these patients. As a result, the biliary drainage catheter is left in the cyst rather than advanced through the long common channel into the duodenum. Patients with choledochal cyst disease may be at special risk of developing pancreatitis because of the high frequency of an anomalous choledochopancreatic duct

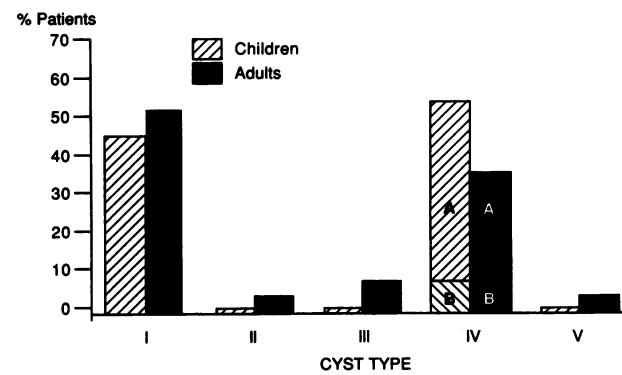

Figure 2. Distribution of choledochal cysts in children and adults. Choledochal cysts were classified based on cholangiography according to Todani.[6]

junction.[12-14] Thus, placement of the catheter into the cyst but not into the duodenum is recommended. Percutaneous cholangiography with catheter insertion primarily has been used in adults, but one of the recent 6-year old children also had successful percutaneous transhepatic drainage. However, in the majority of the young children in this series, choledochal cysts were identified by ultrasonography, and cholangiography was easily performed intraoperatively. Postoperative stenting was required infrequently in these children. In the older children, preoperative cholangiography can be obtained by either the endoscopic or transhepatic approach.

Cholangiography demonstrates and classifies the extent of biliary dilatation. Isolated fusiform dilatation of the extrahepatic biliary tree (Type I) choledochal cysts are the most common in most series,[3,8,9,15] usually accounting for 60% of all cysts. In this series, Type I cysts were identified in 50% of the patients, including 45% of the children and 52% of the adults. As reported by Todani et al.,[6] choledochal cysts involving both the extrahepatic and intrahepatic biliary tree (Type IV) are the second most commonly identifiable cyst type. Type IV cysts also were commonly seen in both children (55%) and adults (35%) in this series. A diverticulum of the extrahepatic biliary tree (Type II) or intraduodenal extrahepatic cystic dilatation (Type III) are uncommon. Caroli's disease isolated intrahepatic biliary dilatation or Type V choledochal cysts, also is uncommon.[16]

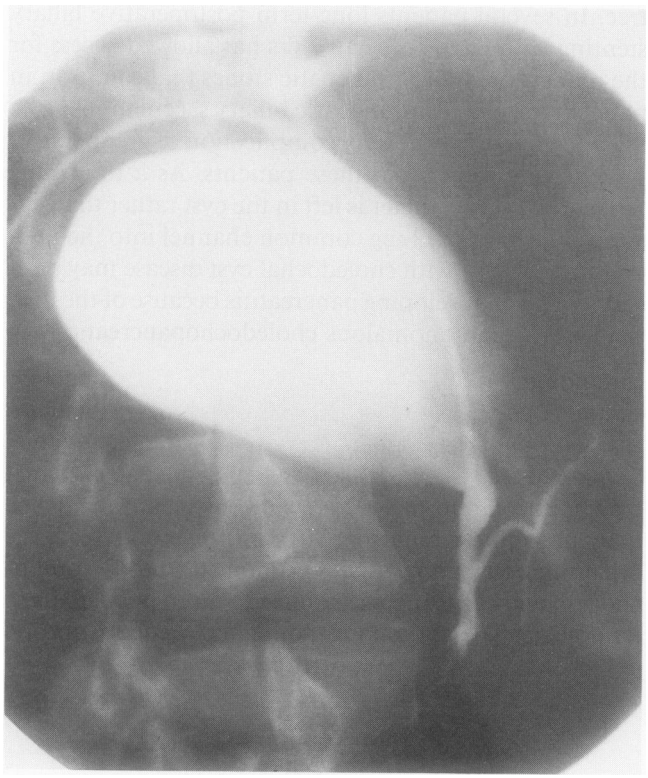

Figure 3. Percutaneous cholangiogram of a young adult with a Type I choledochal cyst. A long common channel of the common bile duct and *pancreatic duct* is easily seen.

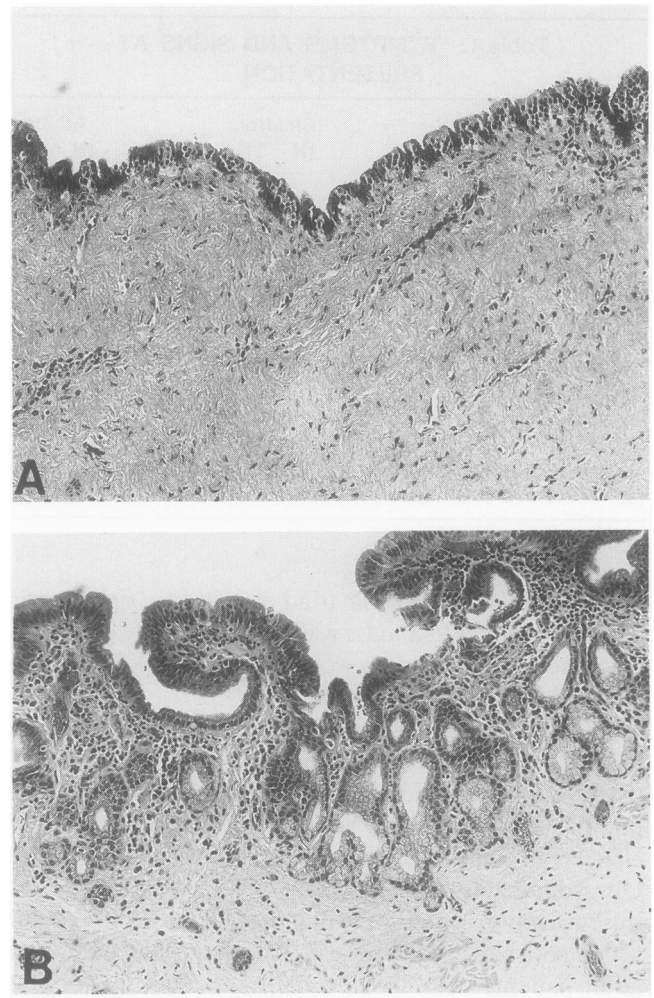

Figure 4. Histologic sections of two patients with a choledochal cyst (A). Densely fibrotic wall of the cyst typically of choledochal cysts in children (B). Glandular type of epithelium seen in an adult with a choledochal cyst. Note inflammatory cells and mucosal erosion (H/E × 40).

The risk of malignant degeneration of choledochal cysts is well documented in the literature.[17-21] The age at diagnosis of a choledochal cyst is related to the later development of carcinoma in the gallbladder, the cyst, or the intrahepatic ducts.[17] In patients who have choledochal cysts at 10 years of age or younger, the risk of developing cholangiocarcinoma is approximately 1%, whereas the risk increases to 15% for patients older than 20 years of age.[17] Not only is age related to the risk of development of carcinoma in a choledochal cyst, but treatment is an important factor. Todani et al.[19] reported a higher incidence (17.5%) of carcinoma arising within choledochal cysts, and this observation was associated with a high incidence of cyst bypass surgery. The fact that

only one gallbladder carcinoma and only two late cholangiocarcinomas were observed in this series probably relates to the high percentage of patients in whom cysts were excised, the relative young age (median 15 months) of the children, and the short follow-up of the adults.

Total excision of gallbladder and choledochal cyst Types I and II is now standard.[4,9,21] Excision is the treatment of choice for several reasons. The previously reported high incidence of postoperative strictures and recurrent cholangitis has improved significantly with this management.[14] In addition, the possibility of carcinoma developing in the cyst or gallbladder is markedly reduced. This series supports the safety of excision and reconstruction over a wide range of ages.

Type IV cysts were common in this series (41%), and total excision of the extrahepatic biliary component is warranted. If associated intrahepatic dilatation is confined to the left hepatic lobe, lobectomy has been proposed by Todani et al.[22] and by Tsuchida.[23] This treatment was required in only one patient in this series. In some circumstances, intrahepatic dilatation has regressed after excision of the extrahepatic cyst component.[24] Excision of the extrahepatic portion is the preferred treatment of Type IV cysts. However, management of the intrahepatic portion of Type IV cysts is more controversial. This group of patients is more likely to harbor intrahepatic stones and have problems with recurrent cholangitis. Thus, percutaneous biliary stents can be helpful to guide the intraoperative placement of large bore transhepatic stones for long-term biliary access. This group of patients should be observed postoperatively for the resolution or development of additional biliary tract problems. In patients with intrahepatic stones or strictures, percutaneous choledochoscopy can be performed easily via the tube tracts for stone removal, balloon dilatation, and surveillance for malignancy.

In this series, choledochal cyst disease was observed more commonly, especially in recent years, in adult patients. However, many of these adult patients had their choledochal cysts identified either after operation for biliary symptomatology or after attacks of pancreatitis. This presentation contrasts with that of children who were identified as neonates, usually with an abdominal mass and jaundice. Preoperative evaluation is similar in children and adults and should consist of ultrasonography or computerized tomography and cholangiography. Some authors have suggested that embryologic biliary obstruction and subsequent common bile duct wall weakening is more important in infants, whereas an anomalous CDPDJ is a important factor in older patients.[13,14,25,26] However, an anomalous choledochopancreaticobiliary junction was seen in both children and adults in this series. Percutaneous cholangiography with drainage is helpful in selected older children and adults. Pathologically, children most often have a dense fibrotic choledochal cyst, whereas adults are more likely to have inflammation and dysplasia without a fibrous component. In addition, reflux of pancreatic juice and bile stasis may contribute to the 5- to 30-fold increased incidence of malignancy in patients with choledochal cyst disease.[18–20] Thus, the pathogenesis of choledochal cyst disease may be different in children and adults. Cholangiocarcinoma is reported to be more frequent when choledochal cysts are identified in adulthood, and this series confirms those reports.

The treatment of choledochal cysts has changed during the last 20 years. In the past, bypass procedures were common; currently, excision of the cyst and hepaticojejunostomy are considered appropriate for most patients. The treatment of choledochal cyst disease is surgical and does not depend on the age of the patient. Excision of the cyst with Roux-en-Y reconstruction of the biliary tree is the preferred treatment in most patients. In patients with Type IV choledochal cysts, cholangitis secondary to intrahepatic stones or strictures may result. Moreover, these patients remain at risk for the development of intrahepatic cholangiocarcinoma, and therefore, they should be observed for follow-up indefinitely.

References

1. Vater A, Ezler CS. Dissertatio de scirrhis viserum occasione sections viri typanite defunte. Wittenburgae 1723: 4 Pamphlers 881, p 22.
2. Worter GL. Congenital cystic dilatation of the common bile duct. Ann Surg 1924; 8:604–626.
3. Yamaguchi M. Congenital choledochal cyst. Analysis of 1,433 patients in the Japanese literature. Am J Surg 1980; 140:653–657.
4. Schmid C, Meyer HJ, Ringe B, et al. Cystic enlargement of extrahepatic bile ducts. Surgery 1993; 114:65–70.
5. Cosettino CM, Luck SR, Roffensperger JG, Reynolds M. Choledochal duct cyst: resection with physiologic resection. Surgery 1992; 112:740–748.
6. Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977; 134:263–269.
7. Deziel D, Rossi RL, Munson L, et al. Management of bile duct cysts in adults. Arch Surg 1986; 121:410–415.
8. Joseph VT. Surgical techniques and long term results in the treatment of choledochal cyst. J Ped Surg 1990; 25:782–787.
9. Lopez RR, Pinson CW, Campbell et al. Variation in management based on type of choledochal cyst. Am J Surg 1991; 161:612–615.
10. Young W, Blane C, White SJ, Polley TZ. Congenital biliary dilatation: a spectrum of disease detailed by ultrasound. Br J Rad 1990; 63:333–336.
11. Componovo E, Buck JL, Drane WE. Scintigraphic features of choledochal cyst. J Nucl Med 1989; 30:622–628.
12. Babbitt DP. Congenital choledochal cysts. New etiological concepts on anomalous relationships of the common bile duct and pancreatic bulb. Ann Radiol 1969; 12:231–240.
13. Okada A, Nakamura T, Higaki J, et al. Congenital dilatation of

the bile duct in 100 instances and its relationship with anomalous junction. Surg Gynecol Obstet 1990; 171:291–298.
14. Rattner DW, Schapiro RH, Warshaw AL. Abnormalities of the pancreatic and biliary ducts in adult patients with choledochal cysts. Arch Surg 1983; 118:1068–1073.
15. Komi N, Tamura T, Miyoshi Y, et al. Nationwide survey of cases of choledochal cysts. Analysis of coexistent anomalies, complications and surgical treatment in 645 cases. Surg Gastroenterol 1984; 3:69–73.
16. Mercadier M, Chigot JP, Clot JP, et al. Caroli's disease. World J Surg 1984; 8:22–29.
17. Voyles CR, Smadja C, Shands WC, Blumgart LH. Carcinoma in choledochal cysts: age-related incidence. Arch Surg 1983; 118: 986–988.
18. Todani T, Tabuchi K, Watanabe Y, Kobuyashi T. Carcinoma arising in the wall of congenital bile duct cysts. Cancer 1979; 44:1134–1141.
19. Todani T, Watanabe Y, Toki A, Urushihara N. Carcinoma related to choledochal cysts with internal drainage operation. Surg Gynecol Obstet 1987; 164:61–64.
20. Kagawa Y, Kashihara S, Kuramoto S, Maetani S. Carcinoma arising in a congenitally dilated biliary tract: report of a case and review of the literature. Gastroenterology 1978; 74:1286–1294.
21. Ohta H, Yamaguchi Y, Yamakawa O, et al. Biliary papillomatosis with the point mutation of K-ras gene arising in congenital choledochal cyst. Gastroenterology 1993; 105:1209–1212.
22. Todani T, Naruse M, Watanabe Y, et al. Management of congenital choledochal cyst with intrahepatic involvement. Ann Surg 1978; 187:272–280.
23. Tsuchida Y, Ishida M. Dilitation of the intrahepatic bile ducts in congenital cystic dilatation of the common bile duct. Surgery 1971; 69:776–781.
24. Todani T, Watanabe Y, Fujii T, et al. Congenital choledochal cyst with intrahepatic involvement. Arch Surg 1984; 119:1038–1043.
25. Iwai N, Yanagihara J, Tokiwa K, et al. Congenital choledochal dilatation with emphasis on pathophysiology of the biliary tract. Ann Surg 1992; 215:27–30.
26. Wong KC, Lister J. Human fetal development of the hepato-pancreatic duct junction—a possible explanation of congenital dilatation of the biliary tract. J Pediatr Surg 1981; 16:139–145.
27. Alonso-Lej F, Rever WB Jr, Pessagno DJ. Congenital choledochal cysts, with a report of 2, and an analysis of 94 cases. Surg Gynecol Obstet 1959; 108:1–30.